Mitraalstenose. De haemodynamische en clinische verschijnselen in verband met de operatieve therapie.

Sien, Que Giok

SUMMARY

Introduction

After a short historical survey of the clinical aspects and of the development of the surgical therapy of mitral stenosis, the subject of this thesis was stated.

Chapter I

In this chapter the etiology and pathogenesis of mitral stenosis are briefly discussed. In nearly 100% of cases of rheumatic fever, the heart is affected. An organic valvular lesion develops in more than half of these patients. Attention is drawn to the occurrence of atypical forms of rheumatic fever.

In the discussion of the pathogenesis, special attention is paid to the recent study of MAGAREY which deals with this subject and to the anatomical types of mitral stenosis described by BROCK, HARKEN and several other authors. These anatomical forms are important in connection with the surgical treatment. Severe forms of mitral stenosis are apparently produced by a slight valvulitis, whereas mitral insufficiency usually is the result of severe damage of the valvular structures.

A fair degree of stenosis raises the pressure in the left auricle and in the pulmonary circulation, which leads to hypertrophy and dilatation of the left auricle and the right ventricle. Secondary changes in the pulmonary vessels and parenchyma may develop due to the increased pressure. These structural changes in the pulmonary vessels, especially in the arterioles, have hemodynamic consequences, and are an important factor in determining the result of operative therapy. Attention is drawn to the bronchopulmonary anastomoses, which play a part in the production of hemoptysis. Finally hemosiderosis is discussed, especially its roentgenologic appearance.

Chapter II

The modern knowledge of the circulatory disturbances in mitral stenosis is discussed in this chapter.

The pulmonary "capillary" pressure ("PC" pressure, PCV pressure, pulmonary artery wedge pressure), determined by the technique of HELLEMS et al., gives an impression of the pressure in the left auricle. The "PC" curve shows cyclic changes, which are believed to reflect the cyclic changes in the left auricular pressure. In severe stenosis (approximately 1 sq. cm) the pulmonary "capillary" pressure is increased at rest. At "PC" pressure exceeding 25 mm Hg pulmonary edema is frequently seen, though exceptions have been described.

The pulmonary artery pressure (PAP) increases when the "PC"

pressure exceeds 15 mm Hg. Initially this increase in PAP is a parallel of the increased PCV pressure. When the PCV pressure is raised to levels higher than 25 mm Hg, a more pronounced increase of the PAP occurs. This is thought to be due to an increase of pulmonary vascular resistance, especially in the pulmonary arterioles. An increased arteriolar resistance is usually found in cases of severe stenosis, though exceptions may be seen. This increased resistance is supposed to be due to anatomical changes in the arterioles, or spasm or both. It is thought to be a compensatory mechanism which protects the pulmonary capillaries from a high hydrostatic pressure and pulmonary edema. The cause of this spasm is essentially unknown. Possibly disturbances of ventilation play a role. In this connection one should mention the observations of McGREGOR et al. who noticed a fall in pulmonary arterial pressure and decrease of pulmonary arteriolar resistance after breathing of 100% oxygen, as well as the experimental work of VON EULER and LILJESTRAND, DIRKEN and HEEMSTRA.

The cardiac output in severe cases of mitral stenosis is frequently subnormal. On exertion it is usually not sufficiently increased and may even show a decrease.

The formulas of GORLIN and GORLIN and DEXTER are briefly discussed. The theoretical objections against them are briefly mentioned. The use of the formulas appear to be useful in clinical practice in judging the severity of the stenosis.

Exertion, emotion and tachycardias may raise the "PC" pressure and consequently produce pulmonary edema. These factors do not influence pulmonary vascular resistance.

The most important disturbances in pulmonary functions in mitral stenosis are a decrease of the vital capacity, impairment of diffusion, and a decreased utilization coefficient of oxygen on exertion.

Chapter III

In this chapter the clinical aspects of mitral stenosis are discussed. This valvular disease is more frequently seen in women. It is especially seen in youth and middle age.

A short survey is given of the different theories about the mechanism of dyspnea in mitral stenosis. Fair to serious exertional dyspnea usually indicates a severe stenosis. This symptom is related to pulmonary engorgement and probably to pulmonary vascular resistance. Patients with a high degree of pulmonary engorgement are orthopneic. Paroxysmal dyspnea also occurs frequently.

Other serious disturbances are cardiac asthma and pulmonary edema, both probably have the same etiology. A strong right ventricle, a high "PC" pressure and reflexes arising from the congested lungs play an important role. The acute pulmonary edema may be the result of acute insufficiency of the left auricle. Cough and recurrent bronchitis as well as hemoptysis are also manifestations of serious mitral stenosis. Hemoptysis may be caused by infarction of the lungs, pulmonary edema and some-

times by rupture of the broncho-pulmonary anastomoses. On the formerly used conservative regime hemoptysis had a serious prognosis. Chest pain, palpitations and fatigue are also observed in mitral stenosis. These symptoms are difficult to explain.

The diagnosis of mitral stenosis is based on the finding of a typical diastolic murmur, a loud apical first sound and often an accentuated pulmonary second sound. The diastolic murmur has to be differentiated from the murmur of aortic insufficiency, relative mitral stenosis, and sometimes from the GRAHAM STEELL murmur.

The opening snap is a pathognomonic sign of mitral stenosis and is thought to be a reliable indication of pliant mitral valves. A loud apical first sound and an opening snap in cases of a combined mitral lesion suggest a predominant stenosis. With severe calcification of the valves the first apical sound is said to be soft. An apical systolic murmur does not necessarily indicate the existence of mitral insufficiency. The blood pressure is most often normal, however it may be elevated especially in middle aged women.

The roentgenologic appearance of the heart may be normal. In many cases a characteristic configuration of the heart is found due to the enlarged left auricle. The right ventricle is often enlarged. Enlargement of the left ventricle in cases of a combined valvular lesion, indicates predominance of the other valve lesion. Calcification of the mitral valves is sometimes seen roentgenologically. The lungs may show the picture of congestion and hemosiderosis.

The electrocardiographic changes most frequently found are changes of the P waves. Electrocardiographic evidence of right ventricular hypertrophy points to a severe stenosis. Signs of left ventricular hypertrophy in a case of a combined lesion indicate predominance of aortic disease or mitral insufficiency.

Phonocardiography usually confirms the physical findings. The opening snap is found by phonocardiography more frequently than it is heard by the observer.

The angiocardiograms of mitral stenosis are characteristic. The pulmonary artery usually is enlarged and remains visible abnormally long. The filling of the left auricle occurs later and also remains visible for a long time. The left ventricle and the aorta are small. Angiocardiography is an important diagnostic method for the evaluation of a concomitant mitral insufficiency, aortic valvular lesions, and a "mute" mitral stenosis.

Electrokymography may give an indication of the degree of a complicating mitral insufficiency.

Sometimes mitral stenosis is found in combination with an atrial septal defect (LUTEMBACHER's syndrome).

Complications which may occur in mitral stenosis are auricular fibrillation, subacute bacterial endocarditis, emboli and infarction of the lungs. Rarely seen are ball-thrombi, paralysis of the left recurrent laryngeal nerve, dysphagia and bronchostenosis.

Chapter IV

The generally accepted indications and contra-indications are discussed in this chapter. Usually patients without complaints are not operated upon. On the other hand, operation should be considered in every patient having symptoms.

Severe exertional dyspnea, paroxysmal dyspnea, pulmonary edema, recurrent bronchitis, hemoptysis, marked pulmonary hypertension and systemic emboli are considered indications for valvulotomy.

Unfavourable features are auricular fibrillation, too small a left auricular appendage, thrombosis of the left auricle, very severe pulmonary hypertension, marked mitral insufficiency or aortic valvular disease, conspicuous calcification of the mitral cusps and functional tricuspid insufficiency.

Absolute contra-indications are intractable cardiac failure, uncontrollable rapid auricular fibrillation, active rheumatic carditis, subacute bacterial endocarditis, predominant mitral insufficiency or aortic valvular lesions, marked tricuspid stenosis and organic tricuspid insufficiency. Most authorities wait at least six months after the cure of bacterial endocarditis and after the rheumatic process has subsided before they submit their patients to operation. The significance of the ASCHOFF bodies in biopsies of the left auricular appendages is not yet clear.

The age limits most frequently accepted are 20 and 50 years, although they are not considered absolute.

The evaluation of the severity of a complicating mitral insufficiency is difficult. One should consider all the available data and should not rely on a single sign or laboratory result. The routine clinical examination supplemented by angiocardiography usually gives a fairly clear picture of the degree of mitral insufficiency.

In judging a concomitant aortic valvular lesion one has to rely especially on the peripheral signs and fluoroscopy. In cases of aortic stenosis the direct arterial tracing (the duration of the systolic upstroke) may be useful.

For the diagnosis of tricuspid valvular lesions, the shape of the right auricular pressure tracings and the height of the right auricular pressure are important supplementary data.

Conspicuous calcification of the valves is an unfavourable feature because of the danger of emboli. Operation in these cases is sometimes impossible and the results are often poor.

Pregnancy is not a contra-indication for operative therapy.

Chapter V

In this chapter the literature concerning the preoperative treatment, the most important complications during the operation, the postoperative course and the results is reviewed.

Preoperatively the aim is to get the patients in optimal condition for valvulotomy. Breathing exercises are considered important in reducing postoperative pulmonary complications. Digitalis is given preoperatively by several authorities with the object of

controlling postoperative auricular fibrillation. This drug is essential in the treatment of congestive cardiac failure and in keeping the rhythm slow in cases with auricular fibrillation.

During anesthesia one should avoid a marked fall of the blood pressure and tachycardia. Adequate oxygen supply should be provided.

Cardiac arrhythmias during operation are not necessarily serious. Active intervention is hardly ever required. Careful selection of the patients reduces the incidence of serious complications like ventricular fibrillation and cardiac arrest. Emboli are apt to occur in patients with a previous history of this symptom, auricular fibrillation or both. The preventive measures are intermittent blocking of the flow through the carotids and the washing out of thrombi which are found in the left auricle. Tearing of the auricular wall and injury of the valvular structures are serious complications.

The postoperative period may be complicated by a clinical syndrome, which is suggestive of a reactivation or a recurrence of rheumatic fever. Pericarditis, arrhythmias especially auricular fibrillation, thrombo-embolic processes, congestive cardiac failure, subacute bacterial endocarditis, atelectasis, bronchopneumonia and pneumothorax are also seen.

The causes of death during the valvulotomy are among others cardiac arrest, ventricular fibrillation, severe hemorrhage from a tear of the auricular wall, cerebral emboli and acute serious mitral regurgitation. In the postoperative period the main causes of death are thrombo-embolic processes and cardiac failure.

The indications which one uses in submitting a patient to valvotomy are a determining factor both for the mortality and the results. The mortality rate varies from 5-10%. Good results are obtained in about 50-60% of cases. Moderate to fair results are seen in about 15-20% and failures in 10-15%.

Factors which unfavourably affect the results of operative therapy, are among others an advanced stage of the disease, a severe accompanying valvular lesion, the surgical anatomy of the valvular structures, an enlarged heart, severe pulmonary hypertension and auricular fibrillation.

A subjective improvement may be accompanied by a reduction of the intensity of the murmurs, the first apical sound and the P_2, but this is not necessary. At roentgenologic examination there is usually no reduction in size of a preoperatively enlarged heart. Several authorities stressed the occurrence of an enlargement of the heart. Also one does not necessarily find improvement in the results of special methods of investigation like electrocardiography and cardiac catheterization in patients in whom the operation has a good clinical result. In the evaluation of the operative results, which is sometimes a difficult matter, these special techniques may provide valuable data supplementing the clinical examination.

Chapter VI

The clinical findings of our patients are discussed in this chapter. They are correlated with laboratory data and with the size of

the mitral orifice as found at the operation. Forty three out of 116 patients admitted with mitral stenosis, with or without an other valvular lesion, were operated upon. The methods of investigation are discussed. These consist of physical examination as well as roentgenologic, electrocardiographic and phonocardiographic examination. Catheterization of the right heart and angiocardiography are applied only when operative treatment is considered. The contra-indications of cardiac catheterization are mentioned. From a total of approximately 1000 cardiac catheterizations in a variety of cardiac and pulmonary diseases, only one lethal complication occurred, resulting from reactivation of the rheumatic process in a patient with mitral stenosis. Two patients developed pulmonary edema from which they recovered.

The total number of 43 patients submitted to operative therapy was made up by nineteen men and twenty-four women. The age varied from thirteen to forty-six years with an average of 30.4 years. Fifteen patients had a previous history of rheumatic fever, one patient had chorea minor. The duration of the symptoms varied from several months to 20 years.

All patients suffered from exertional dyspnea in varying severity, nine patients were orthopneic, six patients had pulmonary edema and eight patients "sub-pulmonary edema". Eight patients suffered from attacks of cardiac asthma, twelve patients complained of recurrent bronchitis. Eleven patients had hemoptysis. Palpitations as well as undue fatigue and chest pain were common. One patient had had systemic emboli, ten patients had suffered from heart failure. The symptoms of exertional dyspnea, orthopnea, pulmonary edema, "sub-pulmonary edema", cardiac asthma, hemoptysis, recurrent bronchitis and congestive heart failure apparently occur in cases of severe stenosis, usually with a mitral orifice smaller than one sq. cm. A significant correlation was not found between the degree of the dyspnea and the mean pulmonary artery pressure (PAP), the size of the heart, the pulmonary arteriolar resistance and the vital capacity. A previous history of pulmonary edema was given by patients with a "PC" pressure exceeding 25 mm Hg as well as by those with pressures lower than this value.

Five patients had auricular fibrillation, four of them had an enlarged heart. Percussion appeared to be insufficient in judging the size of the right ventricle. A presystolic murmur in crescendo was heard seventeen times and an early diastolic murmur in decrescendo twelve times while a combination of these murmurs was found in fourteen cases. An apical systolic murmur was present in ten cases. Mitral insufficiency, which had been predicted after clinical examination was found in five of these ten cases at the operation. The opening snap was present in half of the patients. The apical first sound was in only one case not accentuated. The P_2 was loud in all but four cases.

Chapter VII

At roentgenologic examination enlargement of the right ventricle was found in twelve patients. A double contour at the right

border of the heart, caused by an enlarged left auricle, was present in twelve cases. Two patients had a marked dilatation of the left auricle. The transverse diameter of the heart was enlarged, according to the criteria of UNGERLEIDER and GUBNER. Pulmonary congestion in varying degrees appeared to be present in nearly all cases. There was no distinct correlation between the degree of dyspnea and pulmonary congestion. Hemosiderosis was seen in four cases.

Some cases were described in which the advantages of angiocardiography were demonstrated in the diagnosis and selection for operative therapy. The size of the left auricle as seen on angiocardiograms was very striking if one compares it with the largest diameter of the heart at the same projection. No correlation was found between the mean PAP and the size of the left auricle and with the size of the mitral orifice. There also was no clear relationship between the size of the left auricle and the duration of the symptoms. The changes described by DAVIES et al. in the peripheral pulmonary vessels were observed in fifteen cases. We could not find a correlation between the PAP and these roentgenologic changes.

The main electrocardiographic changes before operation were a right axis deviation in thirteen cases, right ventricular hypertrophy in five cases, abnormal P waves in six and incomplete right bundle-branch-block in one case. The mean PAP in patients with right ventricular hypertrophy showed a fair to strong elevation.

Phonocardiography usually confirmed the findings of the physical examination. The opening snap was present in slightly less than half of the cases. It started usually 0.08-0.12 sec., averaging approximately 0.10 sec., after the maximal vibrations of the second heart sound.

At cardiac catheterization the right auricular pressure was elevated in three cases. One of these patients had clinical signs of a low cardiac reserve. Another patient had LUTEMBACHER's syndrome. The diastolic pressure in the right ventricle was normal in all cases. A normal PAP, which however significantly increased on exertion, was found in only two cases. Six patients had a systolic PAP exceeding 100 mm Hg. An increased PAP was usually found in cases of severe mitral stenosis, but a normal resting pressure did not exclude a severe stenosis. The PAP was increased on exertion in all cases in which it was determined. In about half of our cases this pressure decreased after an intravenous injection of aminophylline. In cases with moderate to severe dyspnea and significant disturbances like pulmonary edema, cardiac asthma, hemoptysis, and congestive heart failure, the mean PAP was definitely raised to levels varying from 41.6 to 54 mm Hg.

The "PC" curves showed cyclic changes resembling those of the phlebogram in all but a few cases. Often significant respiratory changes were seen. In conclusion it can be said that the diagnosis of mitral insufficiency can not be based on the shape of the "PC" curve only, and that this curve does not give a reliable index of the severity of the mitral insufficiency. In cases of a combined mitral lesion, with clear cut signs of insufficiency, the finding of

a "typical insufficiency shape" of the "PC" curve, is however suggestive evidence of the predominance of the mitral insufficiency. In five out of fourteen patients the cardiac output expressed as the cardiac index was below our lowest normal value of 3 l/sq. m/min.

In fifteen out of 37 patients the average PAP-"PC" gradient was lower than the normal upper limit of 10 mm Hg. In severe stenosis the pulmonary vascular resistance is therefore not necessarily increased.

The clinical data of a patient with LUTEMBACHER's syndrome, and one case with a small left auricular appendage were discussed. Attention is drawn to the shape of the heart in the last case in which there was no straightening of the recess on the left border of the cardiac shadow above the left ventricle.

Chapter VIII

In judging a patient for operation we used the indications, discussed in chapter IV, generally accepted by most physicians. Patients with few or no symptoms were not submitted to valvulotomy.

Three patients who were not yet twenty (eighteen, seventeen and thirteen) were operated upon, because they had signs of severe stenosis. In the first two patients severe stenosis was found by the surgeon. In the third case it was not possible to perform valvulotomy because the left auricular appendage was too small. Only five patients were over 40. In our patients over fifty operation was not considered because their general condition was too bad. Some patients with only slight dyspnea were submitted to valvotomy because of other serious disturbances.

Auricular fibrillation was not regarded as a contra-indication, though it is an unfavourable factor. One patient with auricular fibrillation had had multiple systemic emboli which made the operative treatment more urgent.

Five patients were found at operation to have mitral insufficiency as well as mitral stenosis; in two of them the insufficiency was predominant. In these two cases the "PC" curves were "characteristic" of mitral insufficiency. The left ventricle appeared to be enlarged on the angiocardiograms in one patient. By a complete clinical, roentgenologic and angiocardiographic investigation we could with some accuracy assess the degree of a concomitant mitral insufficiency.

Four patients with a complicating aortic insufficiency or stenosis were operated upon because the aortic lesions were not considered serious.

Enlargement of the heart was found in twenty two cases. In our view this is not a contra-indication, except when it reaches an extreme degree.

Heart failure had occurred in ten patients, who were operated upon after the administration of cardiotonics.

One patient was successfully treated for mitral stenosis by operation in the fifth month of pregnancy. The delivery was normal.

Clear cut signs of active rheumatic carditis were absent in our

operated cases. An active rheumatic process we believe to be a definite contra-indication for operation. Microscopic examination of the resected left auricular appendages showed ASCHOFF's nodules in twelve cases. None of these patients had signs or symptoms suggesting rheumatic activity. Only three patients had an abnormal antistreptolysin titer. Three out of these twelve patients possibly have had a reactivation or recurrence of rheumatic fever after the operation. It is possible that a positive biopsy of the left auricular appendage indicates subclinical rheumatic activity, though it has not definitely been established.

Chapter IX

The preoperative care of our patients was discussed. Initially digitalis was administered as a routine measure in a few patients, later on it was used only in cases with heart failure, cardiac asthma, pulmonary congestion and rapid auricular fibrillation.

A serious cardiac arrhythmia during operation occurred only once. In this case it was due to hemorrhage caused by a tear in the left auricular wall. The most important complication during operation was severe hemorrhage which occurred three times. In two cases it was caused by a tear in the left auricular wall, both resulting in death. In three patients the planned valvulotomy was not performed, twice because of predominant insufficiency and in one patient because of marked calcification. Valvotomy was also impossible in a patient with a small left auricular appendage. The size of the mitral orifice varied from 0.3-1.2 sq. cm. In fourteen cases more or less calcification of the valves was found.

In the postoperative period a possible reactivation or recurrence of rheumatic fever was seen in seven cases. Other serious complications were subacute bacterial endocarditis (2 cases), embolism (seven times in the lungs, once in the brain and once in the spleen), cardiac failure (five cases) and shock (two cases). Auricular fibrillation occurred in 14 cases and tachycardia in four. One patient died during this period as a result of shock. In another the cause of death was cardiac failure. One patient died of subacute bacterial endocarditis.

The follow-up period was in the majority of cases more than six months, in some even more than three years. In twenty one patients there was good to very good subjective improvement. In ten cases the results were fair, in seven patients no result was observed. The mortality was 9.5%. A few case histories were discussed as examples of the operative results. In patients over forty years of age the results were definitely worse. We had the impression that the results were not as good in patients with an enlarged heart as in cases with a normal heart. A statistical study however showed no significant difference between the results in these two groups of cases. When the pulmonary artery pressure was very high a good or fair result was also seen in our cases, though some failures occurred. In patients with auricular fibrillation and those with a complicating mitral insufficiency, the results were never very good although some patients showed a fair improvement. Probably this is not caused by the auricular fibril-

lation and the insufficiency per se. We think this is determined by the poor state of the myocardium, and in one patient with mitral insufficiency by associated bronchiectasis. Concomitant aortic lesions which hemodynamically were not important, did not influence the operative results in these patients. A fair to good result was seen in half of the patients with heart failure. In this group however the mortality was relatively higher.

A decrease of the intensity of the diastolic murmur, the first apical sound and the P_2 was observed more frequently in those patients in whom the improvement after valvulotomy was fair to good. An apical systolic murmur developed in a few cases after the operation. The significance of this murmur is not yet clear.

In a few cases a clinical improvement was associated with persistence of electrocardiographic signs of right ventricular hypertrophy and a right axis deviation.

A reduction of the size of the heart was seen in a few patients showing good clinical improvement, whereas enlargement of the heart developed in some patients in whom the operative result was less satisfactory.

A few examples of postoperative cardiac catheterization studies were discussed. We did not observe a parallelism between the clinical improvement and reduction of pulmonary artery and "PC" pressures. In the majority of the patients with a good to excellent result the P. A. and "PC" pressures tended to become normal. In cases with little or no improvement the pressures were still high. A fall of the pressure in the pulmonary circulation shortly after operation in patients who are subjectively improved, definitely indicates improvement of the circulation.

The operative results in our cases are essentially similar to those reported in the literature.

Chapter X

In this chapter our conclusions are summarized. In a given case it is impossible to decide for or against operation on the basis of a single criterion. Only a combination of the subjective complaints, physical examination and special methods of investigation allows us to make a decision. Apart from the physical examination, electrocardiography and roentgenology are valuable. Phonocardiography and angiocardiography are valuable accessory methods of examination, the last one especially in evaluating a concomitant mitral insufficiency or aortic valvular lesion. Cardiac catheterization may also be useful in the assessment of the severity of the mitral stenosis.

Operation is usually not performed in symptomless cases. Severe dyspnea, pulmonary edema, cardiac asthma, recurrent bronchitis, hemoptysis and emboli, and especially combinations of these disturbances indicate the necessity of operative therapy.

Unfavourable features are an enlarged heart, age outside the limits of 20 and 50 years, an associated significant mitral insufficiency or aortic valvular lesion, marked calcification of the valves and very severe pulmonary hypertension.

Absolute contra-indications are: an active rheumatic process, a

subacute bacterial endocarditis, a predominant mitral insufficiency or aortic valvular disease, intractable heart failure, and rapid auricular fibrillation which is not amenable to drug treatment.

During and after operation different complications may occur.

In a given case it is often difficult to get an objective opinion about the result of the operation. The anamnestic data should be evaluated with great care. The result is determined by the indications on which the operation is based.

As far as we can see at this moment valvulotomy is a valuable advance. It is a method of treatment of mitral stenosis which shows great promise.